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SHORT REPORT

A Case of Extensive Retroperitoneal Lymphangiectasia

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Introduction

An unusual case of extensive retroperitoneal lymphangiectasia which presented as an acute abdomen is described.

Case Report

A 21-year-old G6PD deficient male presented just after midnight to the casualty of a community hospital with severe backache, vomiting, a guarded abdomen and a right inguinal scrotal swelling. The swelling had grown slowly over the past 4 years, after an injury to the right testis. An emergency Ultrasound revealed free fluid in the abdomen, a bulky pancreas and saccular dilations in the inguino-scrotal region. The preoperative X-Ray abdomen was unremarkable. The attending general surgical unit performed an exploratory laparotomy.

Free chylous fluid was found in the abdominal cavity. A similar fluid was found in the lesser sac and filling a convoluted mass of massively dilated lymph channels (Fig. 1). This mass was extending from the posterior surface of the duodenum in the retroperitoneal space spreading across the midline. The dilated lymph channels were traced through the inguinal canal into the scrotal sac. There was mesenteric lymphadenopathy. The leaking lymphatic channels were sealed with tie over sutures and the abdomen closed over drains. There was no vascular department in the hospital for intraoperative assistance.

He had a stormy postoperative period. Hypotension followed the 2.5 l loss of chylous fluid. He was transfused with fresh frozen plasma and was on inotropic support. He had rising leukocyte counts, despite higher antibiotics. He gradually settled by the 12th post operative day.

All investigations done in his childhood revealed normal blood counts and the urine showed no chyluria.¹ An MRI² done post-operatively (Fig. 2) showed a large lobulated irregular hyperintense mass in the region of mesentery extending from the level of aortic bifurcation upwards. The mass surrounded the mesenteric vessels. The superior extension of this mass was seen to lie both anteriorly, posteriorly and inferiorly along the pancreas with extension into gastro hepatic recess and retroportal precaval space. The pancreas was pushed anteriorly by this mass. The portal vein was pushed anteriorly and compressed. The coeliac trunk and its branches were stretched. There was evidence of heterogeneous signal intensity

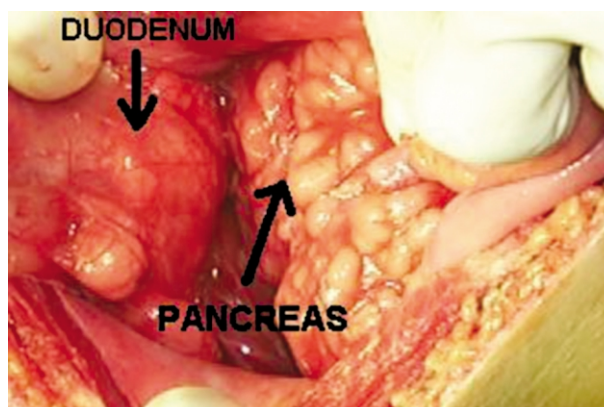


Fig. 1. Dilated lymph channels in the retroduodenal area.

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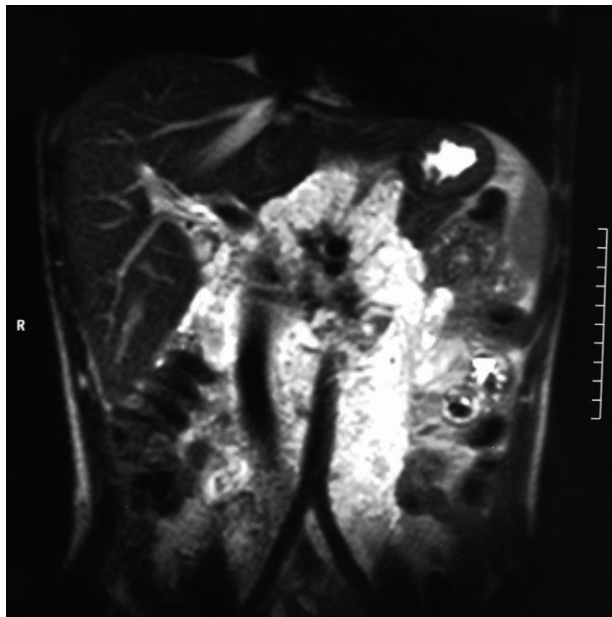


Fig. 2. MRI scan of the patient.

mass in retrocaval region, bilateral paraaortic and prerenal region extending from the level of L1 downwards till the aortic bifurcation. There were extensive preaortic and precaval small soft tissue intensity lesions. Liver, gallbladder, spleen, kidneys and pancreas were normal.

Discussion

Lymphangiectasia or acquired lymphangioma occurs when dilated lymphatic channels arise following

damage to previously normal deep lymphatics. In this case, scrotal injury was, perhaps, a trigger for the lymphangiectasia. There are few cases of Retroperitoneal Lymphangiectasia reported. A review of literature revealed, surgical excision as a cure for symptomatic primary chylous disorders.^{3,4}

The vascular surgeons consulted postoperatively opined that it would have been hazardous to attempt any excision of this extensive lesion during emergency hours. Sealing the leak with abdominal closure over drains was, perhaps, the correct decision for the general surgeon, in the setting of a community hospital. However, in tertiary centres of developed countries, vascular surgeons with acknowledged expertise in dealing with chylous disorders, may consider excision of such a large lesion. Since the exploration, the patient has been admitted once for another episode of severe backache, which was treated conservatively.

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